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Case Report

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Planoepitheliale cell carcinoma arising from ampullo-pancreatic unit

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Summary

Background We report a case of planoepitheliale cell carcinoma located in the ampulla of Vater. We report the surgical results and pathological findings of a patient with an ampullary planoepitheliale cell carcinoma. From what we know it is the first case with successful surgical treatment.

Case Report A 58-year-old woman who presented with jaundice and preoperative endoscopic and radiological procedures could not show whether the tumour was carcinoma planoepitheliale. The clinical presentation, biochemical, radiographic and endoscopic investigation pronounced advanced pancreatic adenocarcinoma.

Results We performed pancreaticoduodenectomy with curative intension. Histological examination revealed: planoepitheliale cell carcinoma of the ampullo-pancreatic unit.

Conclusions In conclusion, preoperative endoscopic and radiological evaluations can all prove insufficient to distinguish between benign and malignant tumour. Thus, for malignant tumours surgeons may be forced to perform extensive surgery.

Key words **periampullary carcinomas • Vater's ampulla • squamous cell carcinoma**

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Background

Squamous cell carcinoma is a very rare form of cancer located in the ampulla of Vater that is derived from ductal cells. Most adenomas and carcinomas of the small intestine and extrahepatic bile ducts arise in the region of Vater's papilla. Histologically, intestinal type adenocarcinoma, pancreatobiliary type adenocarcinoma, undifferentiated carcinomas and unusual types can be differentiated. Histological and immunohistochemical findings support the concept of histogenetically different ampullary carcinomas which develop from intestinal or pancreaticobiliary type of mucosa of Vater's papilla. Molecular alterations in ampullary carcinomas should be correlated closely with the different tumour biology and clinical outcome [1]. As we know, a wide variety of neoplastic lesions may involve the ampulla of Vater, but squamous cell carcinoma has been reported only in one publication from China. The authors reported a case of squamous cell carcinoma of papilla Vater but histological examination suggested a metastatic squamous cell carcinoma. There was only diagnostic process with the end in failure. The patient did not undergo an operation. It is difficult to obtain a histological diagnosis of squamous cell carcinoma of the ampullo-pancreatic unit prior to either operation or autopsy [2].

Aim

We report here the surgical results and pathological findings of a patient with an ampullary planoepitheliale cell carcinoma. From what we know it is the first case with successful surgical treatment.

Case Report

A 58-year-old woman was referred to us with diagnosis of advanced pancreatic cancer. Before that she had suffered from epigastric discomfort, weakness, periodic vomiting, weight loss and 1-year history of diabetes. Physical examination on admission revealed yellowing of the skin and light-coloured stool and there was a palpable firm tender mass deep in the middle upper part of the abdomen. Ascites was absent. The remainder of the physical examination, including pelvic and rectal examination, was negative. The admission laboratory results were as follows: elevated total bilirubin levels (11.01mg/dL), an increase in the white blood cell count to 11,500/mm^3, elevated serum levels of aspartate aminotransferase (161U/L), alanine aminotransferase (420U/L), alkaline phosphatase (521U/L) and elevated serum level of G-glutamyl transpeptidase (794U/L). The serum levels of carbohydrate antigen (CA) 19-9 was elevated (61.63U/mL) and carcinoembryonic antigen (CEA) was normal (1.6ng/mL). A chest X-ray showed no abnormality. Abdominal ultrasonography (USG) and computed tomography (CT) demonstrated intrahepatic biliary dilatation, dilated common bile duct and enlarged gallbladder with a diameter of 110mm. Furthermore, CT and USG revealed a mass in the region of the head of the pancreas with a diameter of 38×37mm. In the vertical part of the duodenum there was stiffening and thickening with 2cm thickness of the wall. A duodenal tumour in combination with a pancreatic one formed an invading mass with a diameter of 60×40mm. An upper endoscopy revealed a duodenal mass with ulceration and marked stenosis in the third portion of the duodenum. There was no possibility to perform endoscopic retrograde pancreatography. An endoscopic forceps biopsy was performed. Histological examination revealed: cellulae carcinomatosae. Clinical data correlated with results of CT, USG and gastrofiberoscopy, and histological findings suggested advanced pancreatic head carcinoma with jaundice and marked stenosis of the duodenum, and the patient was directed for surgery. Consequently, after the patient had agreed, she underwent an operation (13.04.2006). A laparotomy was performed, revealing a well-circumscribed solid tumour, 6 cm in diameter, in the duodenum invading the head of the pancreas without any evidence of extrapancreatic involvement. There was no evidence of metastatic disease in the abdomen. A pancreaticoduodenectomy was performed. The patient had uncomplicated postoperative course and was discharged from hospital 16 days after the operation. Histological examination revealed planoepitheliale cell carcinoma. An immunohistochemical study performed on paraffin-embedded tissue showed positive staining for: cytokeratin high molecular weight (CKHMW), CEA and positive expression of proliferative antigen Ki67 in 30% of cell nuclei. Immunohistochemical characteristics were typical for squamous cell carcinoma. All excised lymph nodes were found negative. No adjuvant therapy was given. The patient was further observed as an ambulatory treated outpatient. She underwent a metastatic work-up. There was no metastatic disease.

Discussion

In the group of patients with periampullary carcinomas treated by pancreaticoduodenectomy,

those with ampullary location are most likely to survive long term. As might be expected, patient outcome has in general been associated with tumour site, stage and type of surgical resection, but some authors have questioned the role of surgical resection for periampullary tumours, concluding that poor outcomes for patients after resection do not justify surgical intervention [3–7]. Periampullary carcinomas (ampullo-pancreatic unit) arise within 2cm of the major duodenal papilla and comprise carcinomas of the ampulla, distal common bile duct, pancreas and duodenum [8]. The most common presenting signs and symptoms include: jaundice 94.4%, weight loss 86%, abdominal pain 64%, nausea/vomiting 47% and pruritus 50% [9]. However, the diagnosis is not easy to confirm before surgery. Endoscopic diagnostics are basic instruments to recognize lesions of Vater's ampulla. ERCP can be a useful diagnostic tool in this setting as it allows inspection of the periampullary region and visualization of the bile and pancreatic ducts. In our case we had no possibility to perform ERCP because of local tumour advance. Menzel et al. showed that overall accuracy for preoperative histopathological diagnosis was 62% with endoscopic forceps biopsies in patients with tumour of ampulla of Vater [10]. In our case we have imprecise preoperative histopathological diagnosis from endoscopic forceps biopsies. Because of the probably advanced stage of the carcinoma there was no necessity for further differentiation of whether the lesion is adeno- or other type of carcinoma. Malignancy arising at the ampulla of Vater is not uncommon and adenocarcinoma accounts for most of these malignancies. Other malignancies of this region have been reported mostly in case reports. In our case the clinical presentation, biochemical, radiographic and endoscopic investigation pronounced advanced pancreatic carcinoma with low possibility of radical treatment. Endoscopic or surgical resection of the lesion is the most effective method of treatment. In the majority of operative cases pancreaticoduodenectomy is performed. In about 80% a curative intended resection is possible [4]. Postoperative radiation therapy with or without chemotherapy does not seem to influence survival in adenocarcinomas, although further study is warranted [11]. We have no information about treatment of squamous cell carcinomas from the literature. There was no recommendation in our case to use chemotherapy. We performed the pancreaticoduodenectomy with curative intension. For resected ampullary adenocarcinoma, several factors have been variably associated with survival: tumour size, histologic differentiation, lymph node status, resection margin status and perioperative blood transfusion [12,13]. In our case there was a large tumour (7×6 cm) with no lymph node metastases and negative margin status. Squamous cell carcinoma is a relatively rare form of carcinoma of the pancreas. Several reports cited an incidence of 0.5–1.9% [14]. We have no information about this kind of histological type of carcinoma arising from the periampullary region except for one case report with probably metastatic squamous cell carcinoma [2]. It is difficult to obtain a histological diagnosis of squamous cell carcinoma of the ampullo-pancreatic unit prior to either operation or autopsy. Several reports about squamous cell carcinoma of the pancreas suggested that hypervascularity may be characteristic in preoperative diagnosis. In our case we did not perform angiography. There was no recommendation to perform it. Planoepitheliale carcinoma is a very rare form of ampullo-pancreatic unit region cancer that is derived from ductal cells of biliary or pancreatic ducts. Basal cells were identified on the basis of immunoexpression of cell-specific markers CK-HMW (cytoplasm). Immunohistochemical study showed positive staining for: cytokeratin high molecular weight (CKHMW), CEA and positive expression of proliferate antigen Ki67 in 30% of cell nuclei in our case.

Surgical resection remains the cornerstone in the treatment of periampullary carcinomas [15]. For all these tumours there is a pressing need for advances in diagnostic capabilities.

Conclusions

In conclusion, preoperative endoscopic and radiological evaluations can all prove insufficient for distinguishing between benign and malignant tumour. Thus, for malignant tumours surgeons may be forced to perform extensive surgery.

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